AUTOPSY ON A CASE OF ATHETOID SPASM MYOTONIA, AND DIFFUSE BILATERAL DISTURBANCES OF SENSATION—CHRONIC CONVEXITY MENINGITIS OF BOTH HEMISPHERES WITH CORTICAL AND SUBCORTICAL SOFTENING—LESIONS MOST MARKED IN THE POSTERIOR PARIETAL REGION.[1]

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THE patient was for more than a year an inmate of the nervous wards of the Philadelphia Hospital, and was the second of two cases used as texts for a clinical lecture delivered at the hospital, and subsequently published in the *International Clinics* for April, 1891. I will summarize chiefly from this published lecture, the most important facts in his history, referring any one desirous of fuller details to the volume. It is important, owing to the post-mortem findings, that an elaborate history of the man had been put upon record.

Nine years before his death he had what was supposed to be a sunstroke, which was soon followed by a fit or seizure, and this by similar attacks every week or two for about a year, and subsequently at rare intervals. He began to develop a spastic state and athetoid movements in the left limbs, probably first in the upper extremity soon after his early seizures, and after the first or second of these his left leg was slightly paralyzed. The following paragraph from the published account, describes his general appearance and condition:

"Inspecting this patient sitting down, it will be noticed that he sits with his head erect, slightly thrown back and stiff, appearing like a soldier in the old fashioned stock—his eyes directed somewhat fixedly to the front. The posterior muscles of the neck, and to a certain extent all of the muscles of this region, seems to be in a more or less spastic state, to which the forced and somewhat rigid position of his head is probably due. Sometimes his head becomes twisted a little to the left and backward. His limbs, trunk and body generally, even when quiet and uninfluenced by voluntary efforts, have more or less spasticity or rigidity. The lines of the forehead are more decided upon the left than upon the right side, as are also the glosso-labial folds of the left side of the face. His mouth is at all times somewhat distorted. The left half of the upper lip has the appearance of being drawn somewhat under, giving the curve of the lip on this side a twisted appearance. His general muscular development is good, perhaps excessive; and the muscles can be seen larger and harder on the left side than on the right."

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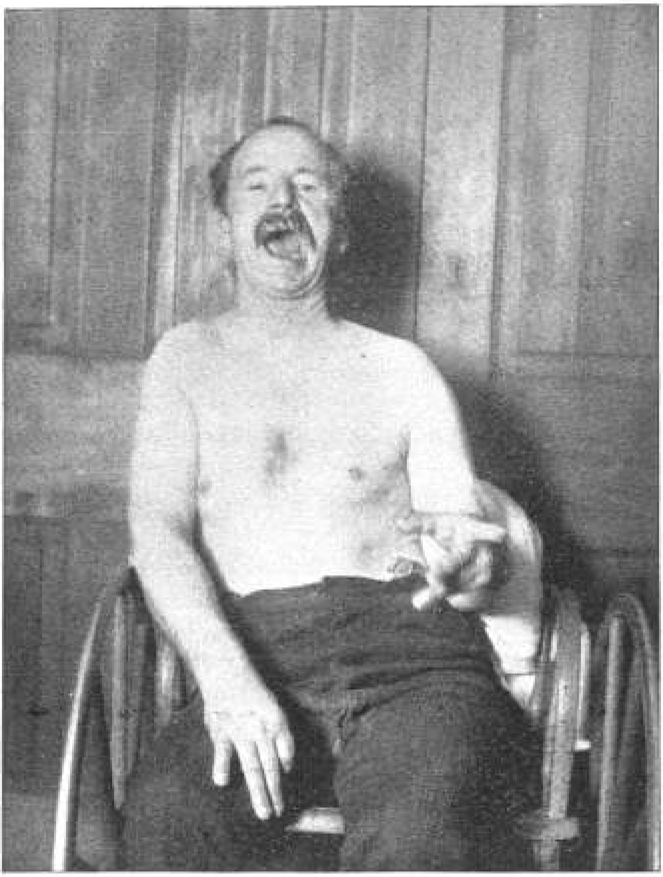
[1] Read before the American Neurological Association, at meeting held at Washington, September 22d, 23d, 24th, 1891.

The most striking feature was certain choreoid and spastic phenomena brought out on any willed action, as opening his mouth, speaking, lifting his hand, etc. The photograph reproduced by permission from the *International Clinics* shows the effects of opening his mouth. The muscles of the face, neck and tongue and extremities took part in a series of athetoid and tonic contractions, much more marked on the left half of the body, although distinctly present on the right. In particular, when he attempted movements with his left upper extremities it was thrown into wild contortions. Manipulation as well as voluntary effort caused the athetoid movements as well as tonic spasms. He presented in brief a curious mixture of rigidity and mobile spasm, usually induced but perhaps sometimes spontaneous. Some paresis of the left leg was present.

The sensory disturbances were reported in the lecture as follows:

" Efforts have been made to test him carefully for sensation, but with results which are contradictory and confusing, owing to the spasm which is induced by the examination, and also perhaps to the general mental condition of the man. Going over him a second and a third time, somewhat different results may be obtained. In general, it was determined that sensation was markedly diminished in the left leg and the outer side of the right thigh, also, apparently, in both feet. A needle could be firmly thrust into the skin in these regions without any response, although



sometimes some reflex muscular contraction was produced. On the inner aspect of the right thigh and leg, less so on the left, sensation to pain seemed to be acute. In testing for sensation he often referred his pain to his toes and jaws, crying out, 'Oh, my toes! oh, my jaws!' accompanying the expression with grinding of his teeth. Over nearly the whole of the left arm and left side of the chest sensation seemed to be lost, deep thrusting of needles giving no result. In the left hand, in one or two places, he seemed to feel; also, at one or two points on the left arm. The dorsum of the hand and fingers, particularly towards the radial side of the arm, was sensitive. On the right side of the chest sensation was acute, repeated, prompt responses being obtained from one prick. These results are not absolutely to be relied on, owing to the answers obtained at different tests. Sometimes he would cry out when he had not been touched. Some of the confusion in obtaining proper responses may have been due to the fact that efforts to fix his attention upon the examination caused the initiation or increase of his painful spastic condition."

Knee jerk was increased and muscle jerk, front tap and ankle clonus were present. Percussion gave phenomena similar to Erb's myotonic reactions.

It is of interest in connection with the findings that he had no headache.

On one occasion, while examining him he had an attack of unilateral Jacksonian epilepsy, chiefly affecting the face and the hand of right side, and associated with marked perversion but not total loss of consciousness. Careful inquiry since the publication of the account of the case, has disclosed the fact that at long intervals he had had similar attacks, varying in severity, and sometimes on one and sometimes on the other side of the body, but oftenest on the left.

The lecture on this patient was delivered about ten months before his death. From that time until his last illness, the spastic, athetoid, sensory, and other phenomena, gradually increased and spread, but not to such an extent as to absolutely disable him.

About midnight, August 24, 1891, he had a convulsion or series of convulsions lasting about half an hour. No record of the character of this convulsion was made. Immediately after it, however, his urine was drawn and was found to contain a large quantity of albumen, and he was treated in accordance with this indication. He remained in bed for three days and was then able to be up and about the ward.

On August 31st he was again taken seriously sick with headache, high fever, and drowsiness. His temperature reached 103°. The spasticity of his limbs and his athetoid movements continued, but not nearly so markedly as when on his feet. Twitching of the right side of his face and of the orbicularis palpebrarum were frequent. He died September 5th; his marked symptoms during his last illness having been headaches, stupor, mental weakness, and fever, with very irregular pulse, respiration and temperature. His urine continued to show a large amount of albumen.

Autopsy.—As I was absent from the city at the time of the patient's death, the autopsy was made by my colleague, Dr. F. X. Dercum, and by Dr. Alfred Stengel, acting pathologist to the hospital; but a few days subsequently I examined the brain. The calvarium was dense and heavy. The dura mater was unusually adherent and nowhere translucent. The pacchionian depressions were not unusual. Over the central part of the lateral surfaces of both hemispheres, that is, chiefly over the superior and inferior parietal gyres, the dura was adherent to the pia mater. The pia was deeply injected, thickened, and infiltrated with plastic lymph, and in numerous places were more or less firm attachments to the brain substance; but the injection, infiltration, exudation and adhesions were much more marked in the postero-parietal regions than elsewhere.

Beneath the area of meningitis, both the cortex and the subcortex were softened, giving the appearance of a sunken subcortical or subpial cyst. On the opposite left side of the brain in a nearly corresponding but somewhat smaller area was a similar belt of inflammation and softening. Subsequent incisions on both sides showed that the softening included the whole of the gray matter, and involved to a considerable extent the white, but did not invade the ganglia or capsules ; it was confined to the supra-ventricular corona radiata.

The vessels at the base were somewhat stiffened, but no other brain lesions were discovered. The pons-oblongata and spinal cord were removed, but no macroscopical lesions could be detected; they were reserved for microscopical examination.

Both lungs were dark and hæmorrhagic, especially at their bases; they contained massive hæmorrhagic infarcts. The heart and liver were normal. The spleen was large and showed two hæmorrhagic infarcts. The right kidney was quite small, measuring only one and one half inch in length, and one and one quarter inch in width; it was markedly lobulated. The left kidney was large and its surface was studded with hæmorrhagic infarcts, several of which had softened and showed purulent walls.

I may be allowed to theorize a little as to the relations of the post-mortem findings to the clinical phenomena.

A convexity meningitis was undoubtedly present, and was so diffiuse as to have possibly caused more or less scattered irritation of the motor cortex. The careful autopsy, however, showed that this meningitis was by far most intense at one particular region, namely, over the superior and inferior parietal gyri in both hemispheres—distinctly back of both ascending convolutions. My own view is that the true motor cortex does not extend back far enough in the human brain to include these two convolutions, but probably ends about the so-called retrocentral convolution, or at the point where the calloso-marginal fissure indents the median edge of the hemisphere.

The choreoid and athetoid phenomena could perhaps be explained by the scattered irritation of the motor cortex; I do not believe that this explanation will hold good for the tonic spasticity and for the sensory disturbances which were present in this case, and we may even perhaps better explain the clonic spasm. The dura mater was so glued to the pia, and the pia-arachnoid so firmly adherent and incorporated with the cortical substance, that many of the blood vessels which form a large part of this inner membrane were obliterated by pressure and increasing inflammation, and it was as a result of this that the cortex and also a considerable extent of the subcortex had necrosed. This softening was on both sides. The subcortical softening was of course chiefly in relation with the same region as the cortical, but from its position it also may have included some of the white motor fasciculi of the corona radiata from the central gyres, and from the gyrus fornicatus and quadrate lobule.

The rigidity or tonic spasticity, as in the other case reported at this meeting, I attribute to the subcortical softening of the motor fasciculi, and it is not improbable that the obliterating of fibres from these motor bundles from time to time, had something to do, in addition to the meningo-cortical irritation with the production of the athetoid phenomena.

From whatever point of view we may regard the case, and I am aware that the effects of the lesions are perhaps capable of somewhat different interpretations, the case is one of importance in the elucidation of several of the still troublesome problems of cerebral localization.

It is, for example, of interest in regard to the localization of the cortical centres for cutaneous sensation. As is well known, at least three views are still held, (1) that the motor and cutaneous sensory centres are identical, or more or less identified—a view ably advocated by Dr. Dana before the American Neurological Association at the meeting of the Congress of 1888; (2) that these centres are distributed in a special and separate region of the cortex, which Ferrier places in the gyrus fornicatus and hippocampal gyrus; (3) that the motor region of the cortex and a separate district of the brain are both concerned in the representation of cutaneous sensations. In my paper on Cerebral Localization, read at the Congress in 1888, I contended for the separate cortical representation of these senses or sensations, and located this sensory zone in a large lobe which includes the hippocampal convolution, the gyrus fornicatus, the quadrate lobule, and posterior parietal convolutions, or at least the upper of these. This lobe is anatomically demarcated by great fissures, and almost enwraps or encircles the motor lobe. I held then and still believe that the sensory cortex is distinct from the motor, but lies nearly everywhere close to it; also that separate centres or areas of representation exist for different zones of the skin, and possibly subcutaneous tissues, as the feet, leg, hands, arms, face, etc.; and that these subdivisions of the general sensory zone are in some way topographically related to the correspoding subdivisions of motor cortical representation.

Because of the difficulties of experimentation, the physiological experiments to determine this localization, while striking have not been entirely satisfactory; embryological, teratological, and anatomical evidences are insufficient; and the clinico-pathological contributions have been conflicting. Dana has made a fair but by no means conclusive showing for the view that the motor and sensory cortex are identical. Certainly if distinct and separate cutaneous sensory centres exist in the cortex, destruction or irritation of them should have some effect; but probably the chief reason that we have so few conclusive cases is that each hemisphere acts for common sensation for both sides of the body to a greater extent than for any other function. Conclusive clinico-pathological observations with reference to this mooted question will be those in which we have autopsies of well studied cases which show corresponding lesions of both hemispheres. Such lesions were presented by this case; lesions which were primarily cortical though to some extent they became also subcortical, as the progress of the meningeal disorder caused obliteration of successive terminal blood vessels. The most extensive and advanced meningeal and cortical lesions in this case were in the postero-parietal region. Here the pia mater was thickened and so adherent to the cortex that it could not be detached, and large but irregularly diffused cortical softening had taken place.

Fortunately, the patient was studied, as carefully as it was possible, for sensation; and although the results above given were not altogether satisfactory they were interesting.

The best explanation of the production of the sensory disturbances in this case would seem to be that they were the result of irritation and destruction of true sensory centres in the postero-parietal region, of commissural association fibres between the sensory and motor cortex; and, thirdly, of fibres ascending from the posterior division of the internal capsule to the cortical sensory lobe which I have described.

The posterior extension of the most positive district of this meningo-cortical lesion was such as to infringe upon and involve the angular convolution. His eyes were examined both by Dr. de Schweinitz and myself, but not for word-vision. It was difficult to make a thorough examination of his vision. Dr. de Schweinitz reported no disease of the fundus oculi except perhaps slightly gray discs. My report states that "he cannot read—that is, he is not able to read because of the manner in which he is affected by his disease. His sight, as near as can be made out, is perfect, but he says he does not read because it hurts him to try to do so. The effort to turn a page, or even to fix his attention upon the reading, gives him pain and throws him into spasms, more or less marked, and of the kind described."

It is not improbable that he suffered from word blindness, but it is perfectly evident that he was in no sense object-blind. His hearing was defective, particularly in the left ear, but he certainly was not word-deaf, although it was necessary to speak to him sharply and loudly in order to make him understand.